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TWO ADDITIONAL CASES OF HEREDITARY CHOREA.

By WHARTON SINKLER, M.D.

TO Huntington has generally been accorded the credit of first describing hereditary chorea,[1] and writers even speak of the affection as "Huntington's chorea," but in Dunglison's Practice of Medicine, second edition, volume second, page 245,[2] will be found a description of what is no doubt the identical disease. The author gives the following letter :—

Franklin, New York, May 5, 1841.

Prof. R. Dunglison :

Dear Sir.—In obedience to your kind request I improve my first leisure since my return home in giving you, in as lucid and satisfactory a manner as possible, an account of a singular affection somewhat common in the southeastern portion of this State, and known among the common people as 'the magrums.' Whence the name originated I do not know, but if it be a corruption of the word 'megrim,' I am at a loss to understand how it ever came to be applied by the vulgar to the disease of which I am speaking, and which has nothing in it analogous to ordinary hemicrania or megrim. It consists essentially in a spasmodic action of all, or nearly all, the voluntary muscles of the system, of involuntary and more or less irregular motions of the extremities, face and trunk. In these involuntary movements the upper part of the air passages occasionally participate, as is witnessed by the 'clucking' sound in the glottis, and in a manifest impediment to the powers of speech. The expression of the countenance and general appearance of the patient are very much such as are described as characteristic of chorea.

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[1] Med. and Surg. Reporter, Phila., Ap. 15th, 1872.

[2] Lea & Blanchard, Phila., 1884.

"The disease is markedly hereditary, and is most common among the lower classes, though cases of it are not unfrequent among those who, by industry and temperance, have raised themselves to a respectable rank in society. These involuntary movements of the face, neck, extremities and body cease entirely during sleep.

"This singular disease rarely, very rarely indeed, makes its appearance before adult life, and attacks after forty-five years of age are also very rare. When once it has appeared, however, it clings to its suffering victim with unrelenting tenacity until death comes to his relief. It very rarely or never ceases while life lasts.

"The first indications of its appearance are spasmodic twitchings of the extremities, generally of the fingers, which gradually extend and involve all the voluntary muscles. This derangement of muscular action is by no means uniform; in some it exists to a greater, in others to a less extent, but in all cases it gradually induces a state of more or less perfect dementia.

"When speaking of the manifestly hereditary nature of the disease, I should perhaps have remarked that I have never known a case of it to occur in a patient, one or both of whose ancestors were not, within the third generation at farthest, the subject of this distressing malady.

"It may not be amiss to state that the last person who came under my observation, and who had the reputation of being an honest man, informed me that, in his own case, this involuntary action of the muscles ceased under the influence of all instrumental music, *except that of the common 'Jew's-harp.'* I very much regret that it was not in my power to test the truth of this statement.

"I also regret that it is not in my power to give any information as to the condition of the catamenia in those laboring under it. I hope to be able to institute a course of inquiry upon this subject during the summer or fall. The disease is markedly hereditary.

"I have thus, dear sir, given you a general, though perhaps no very lucid and satisfactory account of this singular malady. I may observe that, although the description of chorea in the books apply very well to this disease, it nevertheless seems to differ in several respects from ordinary chorea. 1st. It rarely occurs before middle age. 2d. It never ceases spontaneously. 3d. When fully developed it wants the paroxysmal character.

"After all, may not this disease be a peculiar modification of chorea? Is not its pathology in the main the same, and would it not probably be found to yield to the treatment most suited to chorea, if to any. I am, dear sir, respectfully,

Your obedient servant,

C. O. WATERS.

Dr. Clarence King is the only writer I have seen who refers to this communication.[3] I cannot help thinking that the cases referred to by Dr. Waters are in all probability the same which Huntington has reported. The latter says that he, his father and grandfather have known families in Long Island for generations in which this form of chorea has lasted. Dr. Waters speaks of the disease being common in the southeastern portion of the State of New York, and this would correspond with the locality of Long Island, and the character of the cases is remarkably like those of Huntington's.

Another paper on hereditary chorea appeared in 1863, and this, strangely enough, seems to have escaped the attention of Dr. Huntington and the writers on the subject who have followed him. These cases are from New York, and may also be from Long Island, but as no name or addresses are given it is impossible to trace any connection between them.

The paper is as follows:—

CHRONIC HEREDITARY CHOREA.[4]

By IRVING W. LYON, M.D.

"The writer has been familiar from childhood with a type of chorea so unlike in its *origin* to anything described in our standard text books, that the publication of a few facts in relation thereto has been thought advisable, not only as a matter of interest to the reader, but more especially for the purpose of eliciting the observations of any who have met with indications of kindred significance. The peculiarity of origin claimed for this type consists in its hereditary transmissibility; this claim we will endeavor to support by the following facts and considerations.

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[3] N. Y. Med. Journal, Ap., 1885.

[4] American Med. Times, Dec. 19th, 1863.

"The disease, as we have been acoustomed to observe it, has been known in the community by the name of *megrims*. Of the origin or derivation of this term but little can be ascertained, except the conjecture that it may be a corruption of megrain, which word, to say the least, is very inexpressive of any leading character of the malady, which is chorea in toto, consisting of 'irregular action of the voluntary muscles when stimulated by the will,' and marked by an obstinate chronicity. It is confined almost exclusively to certain families, so that such are popularly denominated as "Megrim families," and the children of parents afflicted with this disease are very liable to become the subjects of its manifestation, and in turn transmit it to their offspring. So strong is the conviction of its hereditary influence that the people among whom it occurs believed this to constitute a very legitimate method of propagation, and acting accordingly have repeatedly been known to interdict marriage alliances between their children and those believed to be tainted with the megrim diathesis, under the severe penalties of disinheritance and social ostracism.

"Aged and very intelligent medical gentlemen, who have practiced for the greater part of their professional lives in communities where the so-called "megrims" prevails, testify that they entertain no doubt of its hereditary communicability. We subjoin the histories of three cases.

CASE I.—Mr. A, residing in the town of ——, county of ——, New York, has well marked chorea, which is general, so that he is constantly, when awake, making irregular movements with the upper and lower extremities, facial muscles, and more or less with those of the body. This condition has existed for many years, but seems not to interfere materially with his general health. The digestive function being well performed. Mr. A has two brothers and three sisters; the two brothers have themselves never had any choreal symptoms, but one of them has two children in whom well defined chorea has existed for many years; of three sisters two have had chorea for most of their lives, being now passed middle age. The progenitors of Mr. A on the male side were perfectly free from chorea, but not so on the matenal side—his mother had well developed choreal manifestations from early life, which continued until her decease; she had also a brother who died during adult life from the severity of the disease. But to go still further, both the grandfather and the great grandfather of Mr. A, on the maternal side, had the same disorder which we find in the children. Whether the collateral instance of the affliction occurred in the families we are not advised.

CASE II.—Mrs. K, town of ——, Conn., and a descendant from a family which has long been known and designated as "megrim," had chorea the most of her life, being about seventy-five years old at the date of her death. She had a family of two sons and three daughters; of these one son and two daughters had chorea, with which disease they attained an advanced age. No satisfactory information can be readily obtained in relation to the offspring of the son and one of these daughters so affected; but the other daughter married and had a son, who is now forty years of age, in whom chorea has exhibited itself from puberty.

CASE III.—Mrs. W, formerly a resident of ——, county of ——, New York, had chronic chorea, and lived to an advanced age. She stated that her mother was afflicted with the same disease, together with her mother's father. To these cases many more might be added were it deemed necessary to establish the claim premised."

Huntington's observations, which first attracted the attention of the medical world to hereditary chorea, were published in a paper on "Chorea," which he read before the Meigs and Mason Co.'s Academy of Medicine, Middleport, Ohio, on February 15th, 1872. At the close of the paper he describes a form of chorea, which he says, as far as he knows, is peculiar to the eastern end of Long Island. He, his father and grandfather, whose medical experience had together covered a period of seventy-eight years, had observed for years certain families in which chorea had retained its hold for generations. The chorea presented features which made it distinct and different from chorea as usually met with, and he also mentions that the common form of chorea is extremely rare in that part of Long Island, neither he, his father nor grandfather ever having met with a case there. The features which he particularly points out in the chorea, as seen in the families he describes, are these :—

1st. That it is undoubtedly hereditary. That if a member of one of these families escape the disease the chain is broken and their descendants are free from it.

2d. That the disease is incurable, and that it is likely to be complicated with insanity, a tendency to suicide often occurring in those afflicted with it.

3d. That the disease never occurs before thirty years and seldom after forty.

He mentions two brothers of about fifty years of age, who, although in an advanced stage of the disease and married men, were strongly addicted to flirting with every young woman they would meet, and seemed to see no impropriety in it.

It is to be regretted that Huntington did not give a fuller account of his patients and their family history. It would have added greatly to the value of his paper had he tabulated his cases and traced them back through as many generations as possible.

It is interesting to note that it is in the same part of Long Island where hereditary chorea is found that tetanus prevails to so great an extent.[5]

Since Huntington's observations were published but nine other writers have reported cases in this country and in Europe. The first of these is Landouzy,[6] who reported the case of a man of thirty-seven years of age. His father and one of his sister's died of the same affection, and one other sister had been treated for some "analagous symptoms."

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[5] Hamilton, "Diseases of the Nervous System," p. 375.

[6] Soc de biologie, 1873, quoted by M. Lannois; Revue de med. Aug 10th, 1888, p. 647.

Ewald[7] met with two cases of this variety of chorea.

CASE I.—Mrs. Kratz, aged fifty years, in whom the disease began at thirty-five years. The irregular movements affected the head, face, arms and legs. The speech was clear and easily understood. When at rest the movements were not so great, but when she was excited they increased in intensity. They were controlled to some extent when the patient fixed her attention on any object, as for example in taking a glass of water to the mouth.

The patient's mother suffered from the same disease and died at the age of fifty-eight. She has two brothers and one sister; the brothers are healthy, but the sister, who is fifty-two years of age, has chorea. She has one daughter of thirty years who is so far healthy. The patient has three children who are not yet old enough to have the disease. She knows nothing of her grandparents.

CASE II.—Auguste Otto, aged thirty-seven. She was healthy until thirty years of age when the disease made its appearance. Her grandmother, mother and five uncles died with St. Vitus' dance. She has had five brothers and sisters, of these two are dead and one sister, aged fifty, is afflicted with the same disease.

The next writer on the subject is Clarence King, M.D., who reports a case in an inaugural thesis[8] in 1885. He does not give the locality in which the family lived.

He says: "There has come to my notice a family in which this disease has prevailed for at least four generations, and from which most of the observations of my own are drawn. The first patient known to have had hereditary chorea was the great grandfather of the patients now living.

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[7] Zeitschr. f. Klin. Med., 1884, Bd. vii.

[8] New York Med. Journal, 1885, p. 468.

To him were born ten children, four of whom became sufferers from this disease. Concerning three of this number I am unable to obtain any history further than that they each had children, who finally developed hereditary chorea. Of the fourth child the history is complete. He had nine children, and only one of the number had the disease. Of the eight children who escaped, each has had children, and, although they have all reached the age at which the symptoms usually appear, none of them show any indications of a choreic character. But to the unfortunate one of the family who had the disease five children were born, three of whom were attacked and are the patients now suffering from it. These three patients have no children, and, in all probability, upon their death the disease will become exterminated, at least so far as this branch of the original family is concerned. The two other children are married, but their progeny have not reached the age at which hereditary chorea is developed. Since writing the foregoing I have learned that a brother of the three patients I have cited has begun to shown evidence of chorea. This gentleman, when a child, had the ordinary form of chorea, from which, however, he made a perfect recovery. He is now about thirty-five years of age."

King reported another case[9] in 1886. The patient was a male, aet. fifty-six. He was supposed to be suffering with insanity, but upon examination was found to have hereditary chorea in an advanced stage. The affection had been slowly developing for more than fifteen years. The patient's father died from chorea at sixty-five years of age, having suffered from it for many years. None of the father's brothers or sisters had chorea, but his father died between fifty-five and sixty years of age from the disease. He had been affected for many years. The patient had one half-brother and ten sisters, one sister æt. about sixty has had chorea for many years. The others have escaped, although they have passed the age at which this form of chorea usually begins. The patient is the father of four children, the eldest of whom is twenty-five years.

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[9] Med. Press of Western New York, Dec. 1886, Vol. 1, p. 674.

Perètti,[10] in 1885, reports a very interesting case with a complete table covering four generations. A Mrs. N, whose parents and grandparents suffered partly from chorea and psychical disturbances, had choreic movements. She had four children, two of whom, Anton N and Mrs. A, had chorea. Anton N died at fifty-two, the choreic movements having begun when he was forty. During the last seven years of his life he had disturbance of his mind. He had seven children by his first wife, and three by his second—four children by the first marriage and two by the second had chorea. Mrs. A. died at fifty-seven, and for many years before her death she had choreic movements. She had five children, of whom three became choreic after the age of forty. One of these children married, and had three children, who were living at the ages of forty-seven, forty and thirty-five and were free from the disease.

Huber has written a paper[11] reviewing the subject of hereditary chorea from the time of Huntington's original paper.

Huber's patient was Jacob Rinderknecht, æt 38, silk dyer. The patient was first affected eight years ago; "he awaited his fate with quietness, as he 'knew it was coming.'"

"Status præsens, Oct. 8th, 1886. In bed. Head never at rest. Movements regular. The muscles of expression constantly change. Patient can with difficulty hold his mouth open. Cannot whistle. Speech affected. The patient, when ordered, can make exact movements—as to put a finger on the end of his nose. All the muscles are more or less affected. Sensibility intact. No pain. Urine normal. Can fix eyes and follow the direction of a finger.

Family history—Father died insane at fifty-eight years old, having had the same disease as the patient. There were also affected one brother and one sister of the father, also the patient's grandfather and great grandfather. A sister of the patient's had chorea and was seen Nov. 19th, 1886. Elizabeth Wegman, neé Rinderknecht, æt. forty-two, has a stupid look. Head in constant motion, eyes half shut. In trying to speak, she gives rise to indistinguishable expiratory sounds. Tongue hypertrophied—cannot protrude it. Patient hears well, pupils equal and react to light, gait peculiar, movements cease entirely during sleep. In 1873 she married, and in the same year gave birth to a child. From this time her husband noticed that she was slower in her work and indifferent. In January, 1877, she gave birth to another child, which she did not nurse. She was indifferent, unruly and forgetful. In 1880, when thirty-six years old, her husband first noticed abnormal movements of the elbow, then the arm and leg became affected. In 1881 a change in speech was noticed. In the case of the father of these patients an autopsy was made, and pachymeningitis and internal meningitis were found."

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[10] Berliner Klinische Wochenschrift, No. 50 and 51, 1885.

[11] Virch. Arch. c. viii S. 267, 1887; also Amer. Journal Med. Sc., Oct. 1887.

The "collective investigation committee of the British Medical Association" reports in the British Med. Journal, Feb. 26th, 1887, the following cases observed by Mr. West, of Stoke-on-Trent.

"Charles P., Jr., son of James P., choreic. Stephen, brother of Charles, died at forty; had chorea seven or eight years; was married; his children were unaffected. Charles P., brother of James and uncle of Stephen and Charles, Jr., died of the same disease, aged fifty-one; was affected about ten years. Charles P. had three daughters, one married. Two are quite healthy but very emotional. The youngest is becoming heavy in manner and cross-tempered, just as most of the others who have been affected have begun. She is thirty-three but not choreic at present. The children of the married daughter are healthy. William, a brother of Charles and James, had a daughter who died of this diesase at the age of fifty-six, in America. The father of Charles and James lived to a great age and was quite free. His father lived to over ninety and was free from it. The mother of Charles and James was a Wedgewood, of Etruria, and had it some years before death. It began when she was almost sixty, and was caused by a fright. Other members of her family were given to 'shaking,' and here the first indication ot the hereditary taint seems to have begun."

Mr. West expressed the opinon that the disease was "organic and not the ordinary functional chorea."

In 1884 Mr. West[12] reported the cases of two brothers who had chorea; one died at fifty and the other was still living at forty-five. There was no history of any hereditary transmission in these cases.

McLeod[13] also reported three cases of chorea in adults, two of whom were sisters. There is no reason for considering these cases of hereditary chorea, as there was no family history of the disease, and each of the cases a post-mortem examination revealed a lesion of the motor area of the cerebral hemispheres.

Zacher (Neurolog. Centralbl., 1888, No. 2, and Am. Jour. Med. Sc., April, 1888, reports a case of hereditary chorea.

Patient forty-five years old, well until four years ago, when disturbance of motion began to develop. For two years he had been unable to walk, and had been very excitable and irritable, with a tendency to destroy everything, and was finally committed to an asylum. He exhibits peculiar, irregular, purposeless movements, apparently in all the voluntary muscles. The head is turned from side to side; the face undergoes all sorts of grimaces. Respiration occasionally has a sobbing, rasping sound. The arms are in constant choreiform movement, and the same motion in the legs renders the gait irregular and laborious. The patient is able to control the muscular unrest for a moment and to perform an intended movement with considerable promptness and certainty. He can write his own name well the first time, but a second immediate trial is not successful. The tongue twists about within the mouth and can only be protruded with difficulty. Speech is rather monotonous and with frequent pauses. All movements cease during the sleep. The muscles are well developed and their strength preserved; patella reflex not increased, and sensibility undisturbed. Intelligence seems slight, though it is difficult to determine how far this is due to his imperfect education. The maternal grandfather and two of his brothers had the "shaking disease," while a sister was insane and died in an asylum The grandfather had two sons and one daughter. The latter and one son had the same affection, the other son died away from home and nothing is known of his condition. The daughter had seven children, of whom three are now living and were attacked by the disease after the age of forty; one being the patient under consideration. The patient himself has four children, all young, of whom one is said to have occasional twitchings of the face.

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[12] British Med. Journal, Jan. 5th, 1884.

[13] Journal Mental Science, July, 1881, p. 194.

Various descendants of the brothers of the grandfather are also said to have the same affection, but the author did not succeed in learning anything more definite regarding them. The writer does not refer to Huntington.

Hoffmann (Virchow's Archiv, cxi., 3, 513) reports in full four very interesting cases. Three of these included two brothers and a sister, the fourth being a female cousin. All were attacked about the age of thirty to forty years, except one who developed the disease while still at school, having had epilepsy since two to three years of age. The family history is briefly as follows: Two men of the oldest generation were probably affected by the disease. Next the Grandfather, Waldi, was choreic for several years behis death, at fifty-one to fifty-two. It is not certain whether he was mentally affected. Three of his nine children were choreic. The first was attacked at the age of forty, became weak-minded. One of her two children became choreic at ten, developed imbecility later, and died at nineteen. Another of Waldi's children showed signs of chorea at thirty, and later became insane. She had nine children, of whom three were the first three patients mentioned, and two others were attacked by the same disease at the ages of thirty and forty respectively. A third choreic child of Waldi, a son, exhibited the first symptoms at thirty-eight. There was no mental disturbance (?). His daughter is the fourth of the choreic patients whose case is detailed. The disease therefore extended through four generations and attacked thirteen persons. None of the children of the fifth generation exhibit symptoms, but none are over twenty-four years old. Two of the cases prove that the statement of Huntington is incorrect, that the disease "never begins in youth."

Hoffman then reports another case which developed chorea at forty and also epilepsy at fifty, and whose mother and three sisters had been epileptics. He believes that it is undoubtedly an instance of chronic chorea, and that the absence of psychic disturbance proves nothing to the contrary, since a case of Ewald's exhibited the same peculiarity. As regards the "heredity," it would seem that epilepsy had taken the place of chorea in the predecessors. This epilepsy differed from the usual hereditary form in that it did not develop in youth, but not until twenty-six to fifty years of age. It is interesting to note that in this case chorea followed epilepsy, while in one of the others, as already stated, the order was reversed. The author then discusses the association of chorea with other nervous diseases. Since this case proves an exception to the rule of inheritance, he confers the title "chorea chronica progressiva." He devotes some attention to the possible anatomical seat of the cause of the disease, but sheds no light on the subject.

The most recent case is one reported by Dr. M. Lannois,[14] who has had the kindness to send me his paper, which is by far the most complete and thorough *résumé* of the subject I have seen. Lannois has had the opportunity of studying the cases in six members of the Vey family who were choreic, and he has traced the affection back to the great grandfather.

Jean Sebastian Vey, the great grandfather, had chorea. He had twelve children; of these the history of eight only could be obtained. Five of them had chorea. Joseph Vey, a son of Jean Sebastian, died at fifty-seven years choreic. He had four children, two of whom were choreic. Francois Vey, son of Jean Sebastian, had two children, one of whom was choreic. Catharine, daughter of Jean Sebastian, had five children, four of whom were choreic. The symptoms in the six cases seen by Lannois were studied with great care and correspond with those described by all previous observers.

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[14] Revue de Med., Aug. 10, 1888.

I have had the opportunity of adding two choreic families to those already reported. The first patient came under my care in the nervous wards of the Philadelphia Hospital in March, 1888. The following is his history, for which I am indebted to Dr. Clara T. Dercum, the Resident Physician:

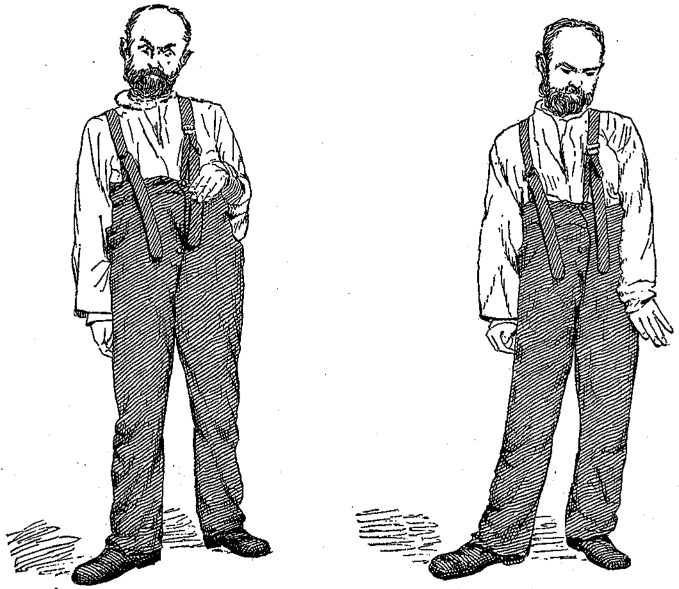
David M., age 43 years; white; born in New York State; residence, Scranton, Pa.; occupation, clerk; weight, 120 pounds; height, 5 feet 3 inches.

The disease with which the patient is afflicted can be traced back to his grandmother's family on his father's side. His grandmother had six children, three sons and three daughters; of these, two sons and one daughter were afflicted. The patient has no brothers and but one sister, who was afflicted in the same manner as himself. She developed the trouble at thirty-five years of age, and finally died of phthisis. In his uncle's the affection began in his legs, his aunt and sisters developed the movements of the head first. In all the cases it took several years for the trunk and all the extremities to become involved. Recently another one of his aunts, who is confined in an asylum in New York State, developed the disease. One of his cousins who is dead had the same affection, although his father, James M., was one of his grandmother's children who escaped. The affection has always been spoken of by the family as St. Vitus' dance. The disease never develops before the thirty-fifth year of age. The father of the patient was accidentally killed at thirty years of age, dying before the age at which the affection appears. His mother died from the effects of a fall. The different members of the family thus afflicted die from some intercurrent affection.

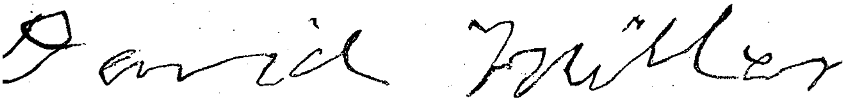
Previous History.—Has had the usual diseases of childhood. Drinks beer moderately. Has had gonorrhœa.

Seven years ago, at thirty-five years of age, patient developed irregular, involuntary movements of the legs. His gait became hesitating and staggering; it gave him no inconvenince, but it annoyed him, because people imagined him drunk. He first noticed it himself in coming down hill, which was only accomplished with great difficulty. Five years ago, at thirty-seven years of age, the same involuntary movements developed in the arms. Two years ago, at forty years of age, the head and trunk became involved.

Status Præsens.—The patient has choreic movements of the whole body, every muscle being involved in these involuntary movements. The head is being constantly turned from side to side, and the arms, especially the left, are continually in motion; the finger movements are somewhat like those seen in athetosis, as shown in cut. The sterno-cleido-mastoid muscles are considerably hypertrophied, although the other muscles of the body, which are also in constant motion, are not enlarged. All the movements are very much increased if patient is watched, or if he attempt to speak or walk. His speech is difficult on



account of incoordination of muscles. His gait is characteristic, he flexes his legs but slightly, plants one foot firmly on the ground, and rests it a moment before lifting up the other foot. There is no motion during sleep, patient lying perfectly quiet. Tongue is protruded easily, but is almost instantly withdrawn with a quick jerky movement. When sitting the patient grasps the side of the chair to steady and rest himself. He has no pain whatever, and is unconscious of the movements himself unless his attention is attracted by others gazing at him. On making a voluntary effort he is able to control the movements to some extent, but cannot stop them entirely. For example, he can steady his hand enough to write his name in a jerky manner.



David Miller

He is able to hold a book so as to read it, and can read aloud in a peculiar spasmodic way, but the book is in constant motion from the movements of his hands, and the head is also shaking, so it is remarkable that he is able to read. He is unable to whistle. He feeds himself with great difficulty. He cannot cut up his food, and in attempting to carry his food to his mouth scatters it about.

Knee-jerk grossly exaggerated. No ankle clonus. Sensation very acute, almost hyperæsthesic. Eyes have been examined by Dr. de Schweinitz, who has pronounced them, as well as the occular muscles, normal in every respect. Examination of the urine revealed it to be normal. All organs have been examined, and as far as can be ascertained are normal. Bowels regular, appetite good, altogether his general health is excellent. His memory is fairly good, and he seems to be intelligent. There are no delusions or evidence of insanity. Table I. gives the family history of James M., as far as can be traced. It was through this patient that I became aware of the other family of hereditary chorea, which I will report later. He told me that at the onset of his troubles he thought that he could keep off the disease by travelling about. He got a peddlar's pack and went about the country selling his goods. In his travels he saw members of a family in Wyoming Co., Pennsylvania, who he thought suffered from the same disease as himself. I wrote to the address which he gave me, and through the kindness of a member of the family who has sent me a most intelligent communication, and Dr. C. R. Newton, of Nicholson, Penna., I have been able to get a very good account of the disease.

The first member of the family of whom anything definite is known was Mrs. D., who was choreic. She had a large family; two of her sons and one daughter were "affiicted." One son never married. Another son had three children; neither of these nor their descendants had the disease. The daughter had two daughters, Elizabeth and Polly; both were "afflicted." Elizabeth married a B., and had six sons

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TABLE I.—M. Family.

The names of those who were choreic are in *italics*.

Grandmother, Mrs. D.

8 children—5 sons, 3 girls.

- 1. Son, James. 7 children.
  - 1. *Son.* Died from heart disease, æt. 36. Slightly affected.
- 2. *Son, Rufus.* Died in insane asylum at Danbury.
  - 2 children. When last seen, oldest one only 30.
- 3. Jesse. 5 children.
- 4. *Son, William.* Was living when last heard from, at Bedford, æt. 70.
- 5. *Son, David.* Died, æt. 30, from a mill accident.
  - 1. *Son David.* Æt. 42; affected since 35. Never married. Our patient.
  - 2. *Daughter.* Died, æt. 39, of phthisis; had been affected about 4 years; had 7 children.
- 6. *Daughter, Mrs. B.* Æt. 70.
  - 4 children. Not diseased when last heard from.
- 7. *Daughter, Mrs. B* Died at White Plains, æt. 65; diseased since 35
  - 2 sons living at Rye, Conn.; 1 daughter died before 30.
- 8. Daughter. Married and had a family. Not affected.

TABLE II.—S. Family. The names of those affected are in *italics*.

Mrs. D. A large family.

- 1. *Son.* Never married.
- 2. *Son.* 3 children. They nor their descendants did not have the disease.
- 3. *Daughter.*
  - 1. *Daughter, Elizabeth* 9 children.
    - 1. *Son* living.
    - 2. Son.
    - 3. Son.
    - 4. Son.
    - 5. Son.
    - 6. Son.
    - 7. *Daughter.*
    - 8. *Daughter.*
    - 9. Daughter.
  - 2. *Daughter, Polly.* 9 children.
    - 1. Burton. Died æt. 2.
    - 2. *Prudence.* 3 children. Afflicted at 30, died æt. 65.
      - 1. *Harmon.* Afflicted at 30; very bad.
    - 3. Dana. Died æt. 61. Parlost his mind. Was not nervous.
    - 4. *Lucy.* 4 children. Afflicted at 40; is now nearly 70.
      - 1. *Milo* (son). Afflicted at 30; now 45.
    - 5. *Nancy.* 3 children. When afflicted, 50; died æt. 56.
      - 1. *Loren.* Afflicted at 40; æt. 45.
    - 6. *Elizabeth* Æt. 67. 10 children. Slightly afflicted.
      - *Several children show symptoms.*
    - 7. *Pauline.* Æt. 64. 3 children. Slightly afflicted.
    - 8. *Perry.* Afflicted at 45; died at 61. 2 children. The one who has just died.
    - 9. Sarah. Died at 2.

and three daughters; one son has been "afflicted," two of the daughters show symptoms. Polly married a S., and had nine children:

1. Burton S., died at the age of two.

2. Prudence S., had three children; she had chorea at thirty, and died at sixty-three. One of her children, H., was "afflicted" at the age of thirty, and has the disease severely.

3. Dana S., died at sixty-one, partially lost his mind, but was not nervous.

4. Lucy S., four children; had chorea at forty, is now seventy. She has one son, M., "afflicted" at thirty, and is now nearly forty-five.

5. Nancy S., had three children; she was afflicted at fifty and died at sixty-six. One of her children, L., had chorea at forty and is now forty-five.

6. Elizabeth S., had ten children, is sixty-seven years of age, and is slightly choreic. Several of her children show symptoms.

7. Paulina S., had three children; aged sixty-four, and is slightly affected.

8. Perry S., had two children; he became choreic at forty-five and died at sixty-one.

9. Sarah S., died at the age of two years.

Dr. Newton, who attended Perry S. in his last illness, wrote me the following letter:

NICHOLSON, PENN., Sept. 14th, 1888.

Dr. SINKLER:

Dear Sir—Yours in reference to the late Perry S. at hand. I saw him two or three days before he died and one or two times previously. This man had chorea for years. Never knew of any cause for the attack, such as excitement, shock, or fright. It seemed in his case to be hereditary, as several members of his family had chorea. He was considered always a clever, energetic man, and apparently inherited no other disease than the chorea. His movements were of the ordinary choreic form, and gradually became worse until he died from exhaustion—failure of heart. The movements ceased during sleep. The intellect was a little impaired for the last few months. There seemed to be a slight paralysis of the muscles of mastication and deglutition and phonation, suggesting possibly the seat of the disease. He complained much for months of "pain and pressure in the occipital region."

Yours very truly,

C. R. Newton.

In reporting these cases I have no new observations to make. I can only confirm and strengthen what has been said by all who have written before me on the subject. It is most remarkable, however, how little variation there is in the family history and symptoms of the different cases reported. All writers have agreed upon the three main points as set forth by Waters and Huntington:

1. The hereditary nature of the disease.
2. That it occurs between the ages of thirty and fifty.
3. That it is incurable, and that there is often mental disease in connection with it.

Huntington and Clarence King assert that if one of a choreic family escape the disease, his descendants also escape it. Although this is usually the case, it is not an unvarying law, as seen in the instance of one of the M. family which, I have reported. James M. did not have the disease, but one son became choreic. In one of Mr. West's cases it is stated that "William P. had one daughter who died of the disease at the age of fifty-six." It is not asserted that William did not have the disease; but as the fact is mentioned in connection with the other brothers, whether they had the disease or not, it is to be presumed that William did not have it.[15]

Another exception to the rule is as to the age at which the affection begins. One of Hoffman's cases[16] developed the disease at ten years, became imbecile, and died at eighteen; and another became choreic "while going to school." One of Lannois' cases had the first symptoms at twenty-one years, and one of Lyon's began "about puberty."

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[15] In the A. family of Lyons there were two sons and three daughters. One son and two daughters had chorea. One son escaped, but his son had it. Walters says the disease may pass over a generation.

[16] Op. cit.

[17] In one of Hubers's cases, Magdelena R., it is not stated that she was choreic, although her son Heinrich had the disease; but this may have been an oversight.

Still these exceptions do not influence the generally observed fact that the disease, as a rule, does not occur before thirty or after fifty years of age.

Sex exerts but little influence on the disease. In the two families which I have reported there are eleven males and fifteen females in a total of twenty-six cases. Lannois gives nine males and ten females out of nineteen cases. One fact of importance is that the disease does not materially shorten life, although eventually most of the patients die as the result of the affection. Still it is notable the advanced age that many of them attain. Among my own cases are the following ages: Prudence S. died æt. sixty-five, Dana S. died æt. sixty-one, Lucy S. living nearly seventy, Nancy died æt. fifty-six, Elizabeth living æt. sixty-seven, Perry S. died æt. sixty-one, M. died of heart disease æt. thirty-six, William M. living æt. seventy, Mrs. B. living æt. seventy, Mrs. B—t died æt. sixty-five.

Another point which strikes one in reviewing the cases reported is the unusual number of large families met with among them. Among my own cases are one family of eight children, two of nine, and one of ten. In the Vey family of Lannois, the great-grandfather had twelve clildren, and Francois, his eighth child, had thirteen.

The character of the movements are markedly like Sydenham's chorea, and I cannot accept the point of difference made by Lannois and others that in hereditary chorea the movements are controlled by effort of will or when a voluntary movement is attempted, for in a certain proportion of cases of ordinary chorea, as seen in children, this is also observed. This has been noted by Mitchell, Gowers, and Hamilton. The movements in the chorea of the aged are quite different from those in hereditary chorea. The irregular motions are never so general or so violent.

The knee-jerk in hereditary chorea seems to be exaggerated. In the case of David M. it is greatly increased, and Lannois found it exaggerated in his six patients. In most of the cases of Sydenham's chorea I have examined, the knee-jerk is lessened or absent, although it is sometimes increased.

We must not fail to note the distinction between hereditary transmission in chorea and the hereditary tendency to chorea. The first is rare, while the latter is common. I have under my care now a girl of six years, who is one of six children, five of whom have had chorea.

There have been but few observations made as to the condition of the heart in hereditary chorea. In the case of David M. there was no evidence of valvular disease, but one of his cousins who had chorea is said to have died of heart disease.

There have been no autopsies made in cases of hereditary chorea which throw any light on the pathological anatomy of the disease. Huber states that the father of his patients died choreic, and at the post-mortem there was found a pachymeningitis and meningitis.[18]

The affection of the intellect and the persistent and gradually increasing movements point to some structural change in the brain, and no doubt in a short time opportunities will be given for determining the lesions in this most obscure and interesting disease.

We cannot but arrive at the conclusion that, although there are many points in common between this chorea and the ordinary form, still there are differences enough to make them distinct and separate affections.[19]

It is more likely that future investigations will show that chorea in the aged is closely allied in its pathological anatomy to the hereditary form of the disease.

I have said nothing as to treatment, because the disease so far has resisted all medication. In the case of David M., hyoscine was administered in large doses, and for a time diminished the severity of the movements. Large doses of arsenic also seemed to lessen them, but there was no material benefit done.

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[18] Dr. William Osler, of the University of Pennsylvania, has recently made a complete autopsy in a well-marked case of hereditary chorea and will shortly publish an account of it.

[19] King's case is of interest in this connection, where the patient had chorea in childhood, made a complete recovery, and late in life had the hereditary form.

One thing we should learn, however, from the study of the cases reported, is that something may be done to prevent the onset of the disease in those predisposed to it by inheritance. When the age at which the disease is likely to come on approaches, the patient should be guarded against all excitement and mental strain; in short, he should be placed under such hygienic conditions as we would think necessary for a patient in danger of mental disease.

In conclusion, I must acknowledge valuable aid in the preparation of this paper to my friend, Dr. H. W. Cattell.